Chondrosarcoma of the maxilla

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SUMMARY

A woman in her 70s reported to the outpatient department of our tertiary care hospital with chief complaints of painless swelling in the right cheek and palatal area. The patient was a known case of diabetes mellitus and hypertension on medications with controlled sugars and blood pressure. The swelling was 10×8 cm in size extending from right infraorbital region up to the lower mandible. CT demonstrated a permeative lytic pattern of bone destruction noted involving the hard palate and maxillary bone.

Using the Weber Ferguson approach, a surgical resection was carried out under general anaesthesia. Resection included from right total maxillectomy (excluding roof of maxilla), nasal septum up to left medial maxillectomy including hard palate and the tumour was resected en bloc. The palatal obturator was fixed. On the basis of histopathology, grade 1 well-differentiated chondrosarcoma was diagnosed. The patient received postoperative radiotherapy and had a good recovery.

BACKGROUND

Chondrosarcomas are malignant tumours with a sluggish growth rate that typically develop from cartilaginous tissues. They usually arise in the pelvis or long bones. Just 5.76% of all chondrosarcoma cases are detected in the head and neck area, where they are infrequently found. In the head and neck region, chondrosarcomas occur more commonly in the maxilla, nasal cavity, nasal septum and mandible.[1] These tumours' genesis is uncertain. They are created, however, from cartilage in tissues that do not typically contain cartilage or, more specifically, from the cartilage cap of an exostosis or an enchondroma. Patients ranging in age from 17 months to 75 years have been documented to develop chondrosarcomas of the head and neck. Incidence peaks between the third and sixth decades of life. They may be well-differentiated growths that resemble benign cartilage tumours or high-grade malignancies that exhibit aggressive local behaviour and the ability to spread. Miles initially described it in 1950. This first report described two cases affecting the maxilla in female patients, both of whom died from the disease.[2]

We present the case of a female patient who reported to our hospital with an aggressive right maxillary tumour. The tumour was resected using Weber Ferguson approach. Histopathology confirmed the clinical diagnosis of well-differentiated chondrosarcoma. An overview of chondrosarcoma along with the clinicopathological findings is discussed in this case report.

This case emphasises the importance of appropriate treatment for good prognosis and we support the view that surgical resection of chondrosarcoma of maxilla with postoperative radiotherapy is the appropriate way of treatment. To determine the prognosis, close and ongoing follow-up are required since tumours having positive or close margins have significantly increased risk of recurrence.

CASE PRESENTATION

A woman in her 70s reported to our tertiary care hospital's outpatient department complaining of a painless swelling in her right cheek and palatal area. Ten months ago, the patient first noticed the bulge, and it gradually progressed to its present size. She had a history of bilateral nasal obstruction, which at first was just localised in the right nasal fossa but later became bilateral. Patient did not give any history of epistaxis, visual disturbance, loosening of teeth. She denied any history of trauma. The patient had a known case of diabetes mellitus and hypertension on medications with controlled sugars and blood pressure. Her general condition was normal. The patient's family history was insignificant. She had undergone previous surgery 10 years ago and had a histopathology report of chondrosarcoma with further details of surgery and grading of tumour not available.

There was evidence of facial asymmetry which was due to the diffuse swelling extending superiorly from the right infra-orbital region up to the lower border of mandible. The swelling extended from midline anteriorly up to the right nasolabial fold and its posterior extent was 2 cm ahead of tragus of right ear. The swelling was around 10×8 cm in size, round, smooth with well-defined margins and had the same colour as the surrounding mucosa. (figure 1).It was non-tender and firm in consistency. The temperature was not raised and skin overlying swelling was not pinchable. No regional lymphadenopathy was observed.

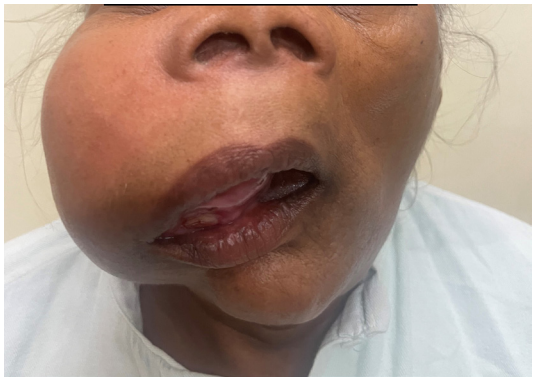

Figure 1 Extraoral profile of the patient showing a large swelling on the right side of the face.

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An irregular, lobulated swelling of about 4×3 cm that covered the whole right side of the palate and had crossed the midline was revealed during an intraoral examination. It was firm, noncompressible, and non-tender to touch, without an erythematous or ulcerated surface. The surrounding mucosa looked normal. The patient was edentulous barring the presence of left upper lateral canine.

INVESTIGATIONS

CT demonstrated a permeative lytic pattern of bone destruction noted involving the hard palate and maxillary bone, predominantly on the right side with evidence of multiple areas of cortical erosions. It was associated with an approximately 7.9×9.2×7.2 cm ill-defined isodense to hyperdense heterogeneously mildly enhancing soft tissue with multiple calcifications. There was no sign of intra-orbital or intracranial extension, and the mass reached superiorly upto the inferior wall of orbit. (figure 2A,B). Ultrasonography showed no evidence of lymph nodal metastasis. Osteosarcoma, chondrosarcoma, Ewing's sarcoma were the differential diagnoses taken into consideration based on clinical and radiological information.

TREATMENT

The patient was hospitalised, and a surgical resection was planned. An impression of palatal defect was taken and a temporary obturator was made by the dental department to cover the defect postoperatively.

Under general anaesthesia, Weber Ferguson technique was used for a surgical resection after obtaining a written, informed and documented consent. Resection included from right total maxillectomy (excluding roof of maxilla), nasal septum up to left medial maxillectomy including hard palate. Soft palate, left alveolus and bilateral floor of orbit were preserved. En bloc resection of the tumour was done after making bony cuts through the right maxillary tuberosity and the hard palate (figure 3). The absence of lymphatic metastasis did not necessitate the need of neck dissection. The palatal obturator was fixed. A liquid paraffin pack was placed in the right nasal cavity and maxillary sinus, removed 5 days postoperatively.

The excision specimen was 10.5×8 cm and had a soft to firm consistency. The majority of areas were soft and gelatinous and had a white grey colour (figure 4). On histopathology, multiple sections examined revealed hyaline cartilaginous matrix with abundant atypical chondrocytes that were organised in lobular patterns. Fibrous bands were seen separating these lobules. The adjacent marrow spaces in the periphery of tumour showed invasive growth of neoplastic lobules. Myxoid degeneration was seen in the enlarged nuclie of cells. No significant necrosis or haemorrhage was seen. Mitotic activity was low to almost absent

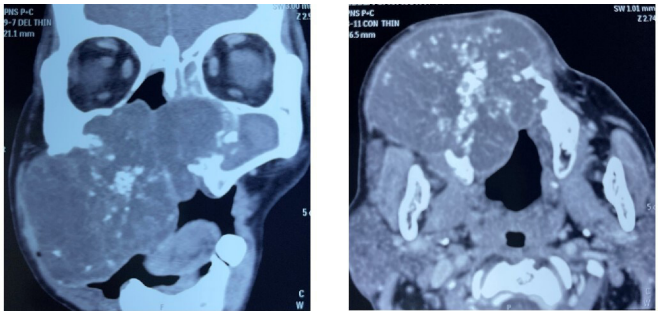

Figure 2 (A, B) CT scan. Coronal section and axial section respectively showing a large isodense to hyperdense heterogeneously mildly enhancing soft tissue with multiple calcifications.

Figure 3 Intraoperative image Weber Ferguson approach.

(figure 5). An impression of grade 1 well-differentiated chondrosarcoma was made. The histopathological surgical margins were tumour-free; however, the right infraorbital and left palatal cut margins were close.

Postoperatively, the patient was started with feeding through a nasogastric tube for few days followed by oral feeding. After undergoing the surgery, the patient was sent for a radiotherapy opinion, which resulted in the patient receiving further radiation treatment on the surgical defect for a total dose of 64 Gy over the course of 3 months following the procedure.

OUTCOME AND FOLLOW-UP

Fifteen days following surgery, the first postoperative check-up was conducted. The cavity after the maxillectomy was in excellent shape. The patient was in fairly good condition at the

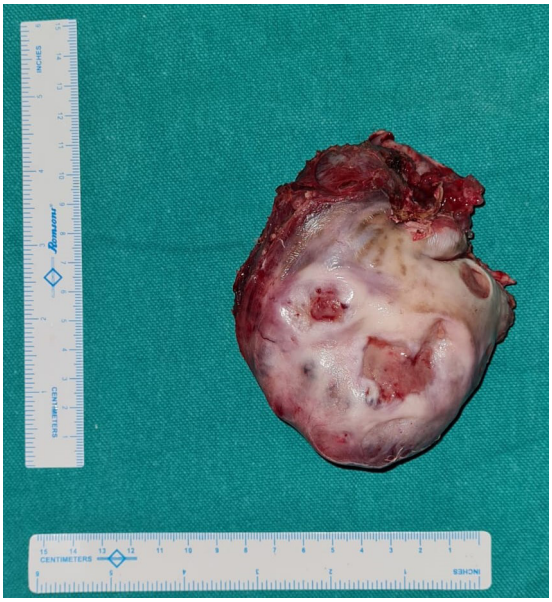

Figure 4 The resected specimen in tot.

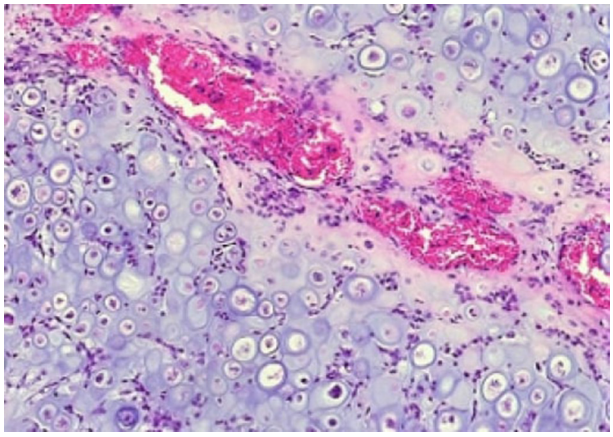

Figure 5 H&E stain.

3-month follow-up, receiving radiation treatments and showing no signs of a local recurrence (figure 6). The patient was irradiated. The patient has been seen for consultations at a rate of every 3 months up to now, and follow-up examination will be done on our patient on a long-term, regular basis at a rate of every 6 months from now until 5 years from now, after which they will be done once a year. Clinical examination and, if necessary, CT imaging will serve as the basis.

DISCUSSION

The maxillofacial chondrosarcoma is quite uncommon, according to study of the literature. They comprise about 10% of all primary tumours of skeleton but are considered to rarely involve the jaws.[3] The average patient age in head and neck cases is between 35 and 45 years old. The most frequent predisposing conditions include multiple hereditary exostosis, Ollier's disease, and Maffucci's syndrome. Patients with Maffucci's syndrome and Ollier's disease have a 25%–30% risk of developing chondrosarcoma with these patients being generally younger than those with primary chondrosarcoma.[4] The neoplasm has a very poor long-term prognosis and exhibits aggressive local behaviour as well as a significant potential for metastasis. This tumour is distinct in that it may develop in either bone or soft tissue, with a roughly two-to-one preference for skeletal origin. The Sinonasal tract chondrosarcoma is quite uncommon. Initial symptoms that a patient may experience include nasal obstruction or discharge, epistaxis, headaches, facial asymmetry, restricted ocular movement, proptosis, diplopia, facial pain, loss of dentition or nasal/aural fullness.

Chondrosarcomas are graded on a 1–3 scale corresponding to a histological classification from a well-differentiated to undifferentiated tumour, based on the rate of mitoses, cellularity and nuclear size.[5] In grade 1 chondrosarcomas, there is an abundance of chondroid matrix along with clusters of chondrocytes that have normal or slightly enlarged nuclei, few nucleoli, no mitoses, and sometimes binucleation, as in our case.[5] Compared with grade 1 tumours, grade 2 chondrosarcomas seem to contain more chondrocytes and less chondroid matrix, infrequent mitoses, multinucleation and somewhat enlarged vesicular hyperchromatic nuclei. Grade 3 chondrosarcomas display a myxoid matrix with irregularly shaped chondrocytes and increased cellularity, nuclear pleomorphism, and mitoses compared with the low-grade chondrosarcomas.[5] In chondrosarcoma, bone formation takes place on framework of pre-existing cartilage matrix; whereas in osteosarcoma, it is directly by the malignant stromal cells.[6] Enchondroma's histological appearance may resemble

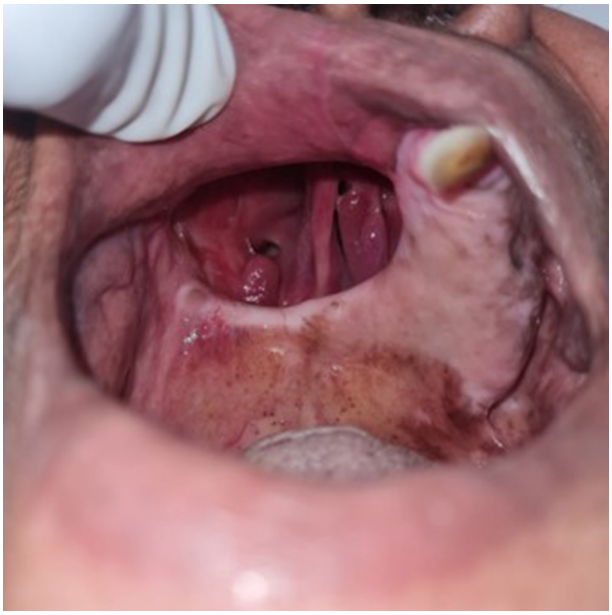

Figure 6 Postoperative follow-up palatal image without obturator.

grade 1 chondrosarcoma. Both calcification and ossification are typical in these situations. Chondrosarcoma, however, exhibits substantial hypercellularity and atypia in higher stages.

Maxillary sinus chondrosarcoma needs to be differentiated from maxillary sinus cancer, maxillary sinus osteosarcoma, fibrous dysplasia, ossifying fibroma, maxillary sinus hemangioma with calcification and maxillary sinus fungal infection. Maxillary sinus cancer is more common in middle-aged and elderly people and typically presents as an irregular soft tissue mass with osteolytic bone destruction.[3] Calcification in maxillary sinus cancer is rare. When the tumour invades the bone of the sinus wall, the signal of the fat band outside the sinus wall is abnormal or interrupted, and this feature is of great significance for differential diagnosis. On CT imaging, the most prominent features of maxillary sinus osteosarcoma are osteolysis and/or osteoblastic destruction as well as an uneven tumour margin. Benign bone lesions like ossifying fibromas (OF) of the maxillary sinus are most common in young children. CT shows that the tumour lesions grow expansively around the medullary cavity as the centre, the sinus cavity is deformed but the boundary is clear. The adjacent structures are compressed but not destroyed. A skeletal developmental defect known as fibrous dysplasia (FD) causes the normal medullary bone to be replaced by fibrous tissue. The lesion grows diffusely and merges with the surrounding bone, with ill-defined borders, and the involved bone expand with a ground glass appearance. Maxillary sinus hemangioma is a vascular tumour, and a small number of cases may appear with small pieces of calcification or phleboliths. In elderly people, maxillary sinus fungal balls (MSFBs) are more common. According to the literature, they show a higher frequency of calcification and partial opacification with an irregular surface on CT scans compared with maxillary sinusitis and other maxillary sinus lesions.[7]

Due to the varying tumour components and the possibility that certain biopsy specimens may only have one of the two parts, diagnosis of biopsy specimens is still difficult. In one clinical series, only 38% of cases were accurately diagnosed on initial analysis.[8]

The procedure of choice, radical surgical excision, has been shown to have the highest likelihood of producing a cure.

Nevertheless, due to the complex anatomy of the maxillary bone area, chondrosarcoma surgical excision with an appropriate margin may be difficult. Recurrences have been linked to surgical margins that are insufficient or incomplete. The overall recurrence rate within a 10-year duration was 7.25% (29/400 patients) in a large series studied.[9] Metastatic neck disease on presentation has been reported to be extremely rare in several studies.[3] In a review of 400 cases of head and neck chondrosarcomas, only 5.6% presented with regional metastatic disease and 6.7% had distant metastasis.[9]

The dual functional and aesthetic functions of the midface's three-dimensional architecture makes reconstruction of the maxillary and palatal deformity quite challenging. The patient can be advised either a prosthetic rehabilitation or a cosmetic reconstruction with secondary flap. The utility of prostheses is limited by their pain when worn, removed, cleaned, and they need to be readjusted often. Nonetheless, prostheses are a successful and cost-effective way to restore speech and mastication. Flap reconstruction involves a longer recovery time with increased risk of surgical complications.[10] There is a wide variety of treatment techniques available to properly repair minor to moderate palatal abnormalities. In spite of the fact that free flaps are used more often, a temporalis muscle flap could be used instead. Bone reconstruction has to be taken into account for medium- to large-sized maxillectomy defects. The subscapular system of flaps, the fibula, osteocutaneous radial forearm and the iliac crest flaps are the composite free tissue flaps that are most often employed in the repair of bone and soft tissue in the maxilla and palate. Studies that compare prosthetic obturation with reconstruction of a palatomaxillary defect demonstrate that there are some advantages to reconstruction, in particular, quality-of-life issues including comfort, convenience, and decreased feelings of self-consciousness.[10] With respect to speech and swallowing function, palatomaxillary rehabilitation outcomes between prosthetic intervention are comparable flap reconstructions.[11] In our case, rehabilitation with a dental obturator was done considering the size of the palatal defect, preservation of the orbital floor, the socioeconomic status and the age of the patient.

The use of postoperative radiotherapy for high-grade or extensive lesions has not routinely been reported; however, chondrosarcomas are susceptible to irradiation and are potentially radiocurable.[12] There are no reports of successful adjuvant chemotherapy for head and neck chondrosarcomas in the literature. Chondrosarcomas, which are traditionally thought of as radioresistant tumours because to their slow response to radiation, do not often receive postoperative, adjuvant radiation treatment. However, the utility of postoperative radiotherapy in cases of unresectable disease or inadequate surgical margins is suggested.[13] The total dose of radiation ranged from 40 to 70 Gy delivered in 30–35 fractions, which is comparable with treatment regimens used for other sarcomas.[3] Krochak *et al* reported survival at 5 years for 38 patients who underwent radical radiotherapy.[14] Thirteen of 25 patients with favourable features were progression-free at the 4-year follow-up, which led the authors to conclude that chondrosarcoma might not be radioresistant.[14]

Patients who have chondrosarcoma have a prognosis that may be affected by a number of factors, including the location of the tumour, whether or not the first surgical resection was successful, and the histological grade of the neoplasm. If the tumour cannot be removed with adequate margins, there is a considerable risk of recurrence, progression and potential metastasis. With chondrosarcoma, the most common reason for death is not metastasis but rather a recurrence of the cancer at the original site. The resection of the affected region in its entirety is of the utmost importance for achieving the effective results.[15] Centrally occurring tumours and the ones with higher grade of differentiation have poorer prognosis. A review of the American College of Surgeons' national cancer database of 179 head and neck chondrosarcomas cases showed a disease-specific survival of 87.2% at 5 years and 70.6% at 10 years.[9] Intracranial tumour invasion frequently causes death in people with head and neck chondrosarcoma.

Although our case was grade 1 well-differentiated chondrosarcoma on histology, with free surgical margins, a portion of the specimen had close margins, necessitating postoperative radiation with 64 Gy to treat the surgical defect. Two years following the surgery and radiation treatment, there was no sign of a local recurrence or distant metastases.

Learning points

- In the treatment of head and neck chondrosarcomas, broad surgical resection followed by postoperative radiation therapy in unresectable tumours and tumours with close margins is crucial. This case and literature review continue to support this idea.
- This rare entity needs to be considered by pathologists and clinicians in the differential diagnosis of neoplasm in the maxillofacial region.
- There is scope for more research in the treatment and reduction in recurrence of patients with this rare neoplasm.

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